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Case Report

Giant mesenteric fibromatosis – A case report

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Abstract

Mesenteric fibromatosis or intra-abdominal desmoids tumor is a rare clinical entity. It is a locally aggressive tumor but not metastasizing proliferation. We report a case of a 27-year-old woman with no significant medical and a previous cesarean section, admitted for abdominal pain.

The CT abdominal evaluation revealed subserosal uterine fibroid. The patient underwent an elective laparotomy and a mass measuring 20/15/15 cm in diameter to the medial wall of the cecum was discovered. Complete removal was accomplished with right hemicolectomy.

Histopathological examination reported mesenteric fibromatosis. Postoperatively, patient was well and 6 month follow-up showed good recovery. Moreover the next month she became pregnant and the pregnancy and birth were uneventful.

Keywords: mesenteric fibromatosis, abdominal desmoids

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Introduction

We present a case of a 27 year old female patient, who was admitted for mid-abdominal pain for about four months. She also had a cesarean section 8 months before and trauma intervention in the past, no taking any medication, no history of smoking. Abdominal examination revealed a mobile firm and globular mass without tenderness located in the lower abdomen. Blood parameters were within normal range and the tumor markers results were negative.

She underwent gynecological exam and transvaginal ultrasound scan that could not establish the belonging of the lump to the uterus or ovaries. Exploratory laparotomy was indicated. Preoperative computer tomography scan revealed a voluminous mass of 124/90 mm base implantation on the uterine serous (Figure 1). After preoperative workup, under general anesthesia, a laparotomy was performed which revealed a 20/15/15 voluminous tumor formation belonging medial wall of the cecum, displacing the right ovary and the uterus to the left.

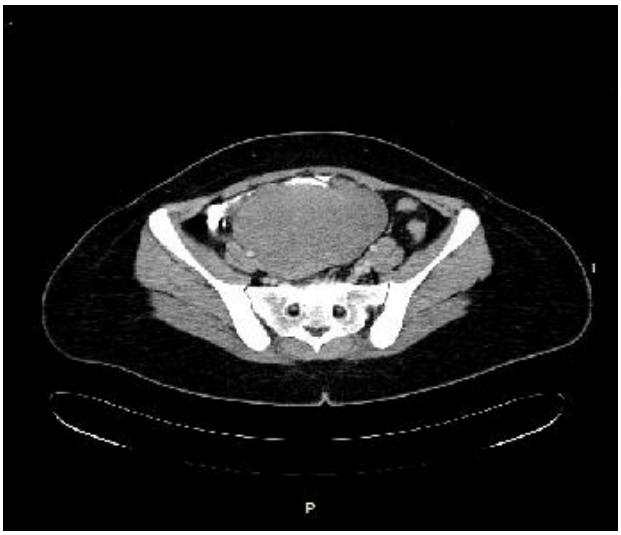

Figure 1. Contrast enhanced computed tomography scan showing a huge inhomogenous mass occupying the pelvis

The mass was excised, performing in addition a right hemicolectomy (Figure 2). The postoperative course was uneventful, no postoperative complications were noticed. The histological diagnosis of mesenteric fibromatosis was determined without any infiltration of the bowel (Figure 3). Six months after resection, a CT scan of the abdomen showed no evidence of residual or recurrent tumor. Shortly after CT scan the patient became pregnant and the pregnancy was uneventful. She had a second CS without any complication apart of the important adherence syndrome.

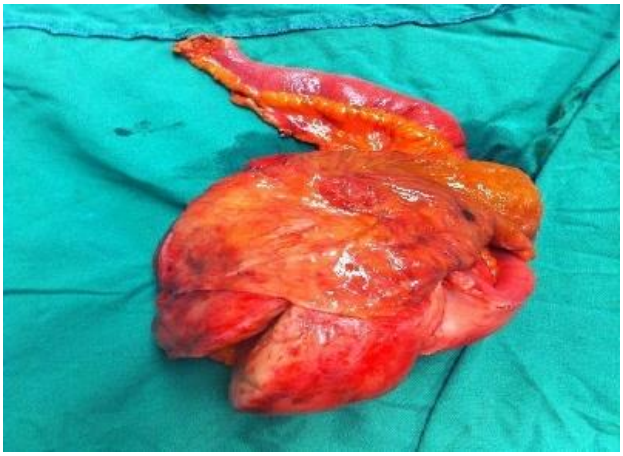

Figure 2. Tumor after resection with attached segment of the bowel

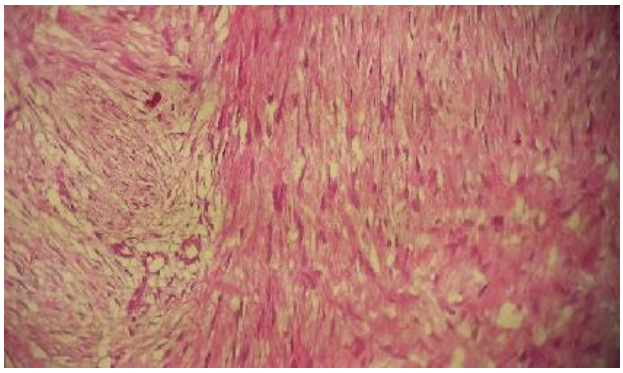

Figure 3. Hystological aspect of the tumor; stellate and spindle cells arranged in a storiform pattern

Discussion

Fibroids can affect both the superficial and the deeper parts of the body. Superficial fibromatosis involves the face, neck, hands, legs, thigh and torso (1). The deep variant affects abdominal wall, mesentery, retroperitoneum and abdomen (2). Fibromatosis is rare, accounting for 0.03% of all tumors. Damage mesentery is very rare. In many cases, tumors are related to Gardner's syndrome and occur most often in young women during or after pregnancy. Patients with a family adenomatous polyposis (FAP,

Gardner's syndrome) have predisposition to the development of mesenteric fibromatosis. Desmoid tumors develop in about 10% of patients with FAP and most are intra-abdominal. Mesenteric fibromatosis associated with FAP seems to follow a more aggressive course and recurrence after resection is high. The etiology is unknown, but an endocrine cause is suggested by prevalence in the perinatal period, tumor regression after menopause or after treatment with tamoxifen (3-5).

Pathogenesis of fibromatosis was unclear for several years. Particularly postoperative injuries can contribute to the formation of these lesions. The growth of such tumors is the slowest in young girls and menopausal peaks, which lead that estrogen may act as a growth factor (6, 7). Currently, these tumors are considered to be a clonally proliferation of myofibroblasts showing APC (adenomatous polyposis coli) gene mutations. These mutations lead to beta-catenin expression (8, 9).

Mesenteric fibromatosis is a rare entity with no clear data for incidence and characteristics. Mesenteric fibromatosis is most common in young adults, with a peak incidence at 30 years with a female predominance (10). Of note, according to literature data, C-reactive protein is significantly increased and this could be induced by inflammatory factors produced by the tumor, such as those produced in inflammatory fibrous histiocytoma (11).

A preoperative differential diagnosis of mesenteric fibromatosis may include intestinal carcinoma, carcinoid tumors, lymphoma and retroperitoneal fibrosis. In our case the differential diagnosis should include uterine myoma, solid ovarian tumor and iatrogenic foreign body. Since there is no classical symptoms related to the mesenteric fibromatosis, the diagnosis is confirmed only after the histological analysis of the tumor (12).

Most patients with mesenteric fibromatosis are clinically asymptomatic. The patient may seek medical advice because of a palpable mass or abdominal pain, or to come to clinical attention because of complications such as gastrointestinal bleeding, bowel obstruction, intestinal perforation or fistula formation. Desmoids diagnoses are usually larger than 5 cm, and can be more than 15 cm. In 10-15% of cases, can be multiple (13, 14).

Imaging remains the gold standard of preoperative investigations to establish a diagnosis of mesenteric fibromatosis work. Radiographs may present a soft tissue mass. Barium studies may reveal compression and angulation of the distal duodenum, the separation of small bowel loops and, more rarely, mucosal tethering (15). The sonographic features of mesenteric fibromatosis are nonspecific and depend mainly by collagen, fibroblasts content and lesional vascularity of the tumor (16). Due to the aggressive tendency to invade adjacent structures, CT scanning is considered first-line modality for identification, characterization and staging fibromatosis. These tumors appear as a soft tissue mass displacement/ viscera involving around usually appear as packing intestinal loops. MRI reveals these tumors hypo-intense on T1-weighted images, due to their major fiber composition (17, 18).

Mesenteric fibromatosis can measure 5-10 cm size on average, although larger injuries up to 24 cm were reported (19). Microscopically, mesenteric fibromatosis is characterized by a proliferation of spindle cells consistent corrugated space without atypia, associated with collagen between dilated vessels. Mitotic count is low, without evidence necrosis and nuclear dedifferention. Immunohistochemistry demonstrates the tumor cells were positive for vimentin and smooth muscle actin. The differential diagnosis includes reactive histological fibrosis and fibrosarcoma. The absence of pleomorphism, mitotic activity significant or atypical

mitosis helps to distinguish fibromatosis fibrosarcoma (20-22).

The management of mesenteric fibromatosis can be difficult. Although they are benign, they can become large and so there is a significant morbidity associated with surgical resection; however, there is no other definitive treatment available. Wide surgical excision is the treatment of first line mesenteric fibromatosis. As mentioned in our case, most of these injuries require bowel resection of the attached segment (23, 24). Radiation may be used before surgery in case of recurrence or inoperable lesions. Mesenteric fibromatosis adjuvant radiotherapy reduces recurrence in 20%-40% compared to 40%-70% with resection only. In cases where surgery and radiation therapy are not satisfactory, systemic therapy with pharmacological agents (antiproliferative and cytotoxic drugs) may be used, including antagonist of estrogen tamoxifen, nonsteroidal anti-inflammatory agent drug sulindac and chemotherapy dactinomycin, vincristine and cyclophosphamide. Surgical excision is the gold standard primary treatment for mesenteric fibromatosis (24-26).

Conclusions

There was not established a protocol optimal treatment for desmoid tumors, but in most cases require a multidisciplinary approach, including surgery, chemotherapy and radiotherapy. Several new approaches to pharmacological and biological treatment are actively developing, although it need long-term follow-up for their foundation. The importance of accurate characterization of a tumor infiltration in the intestinal wall and the mesentery to plan appropriate treatment because the tumor diagnostics based on immunohistochemical staining or traditional histological criteria alone are not specific enough.

We presented a rare case of a mesenteric fibroid tumor imposing differential diagnosis with genital solid tumor, with an excellent outcome after surgery.

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